EXTRA-PAPILLARY COLOBOMATA

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(With eleven wood-cuts and eight chromo-lithographic plates.)

I HAVE chosen the name *extra-papillary colobomata* to signify all those colobomata which do not involve the optic disc, and which are therefore distinct from the classical coloboma due to the imperfect closure of the fœtal cleft, as well as the coloboma of the sheath of the optic nerve, so fully described by Fuchs, van Duyse, and others. I wish to avoid the term central or macula coloboma, because I think I am in a position to show that extra-papillary colobomata, identical with those described by previous writers as peculiar to the macula lutea, may be found in any part of the fundus whatsoever, and I think the reason why they have hitherto been found only about the macula and the disc is, that these are the two parts which form the chief features of interest in the fundus, and are therefore those most carefully examined. Moreover, colobomata unconnected with the disc or macula occur for the most part in absolutely healthy eyes possessing normal vision, and have consequently been overlooked. The so-called macula coloboma must, therefore, be included in, and considered merely a variety of, the extra-papillary colobomata I am about to describe.

Several writers (v. Ammon, Brücke, Huschke, Manz[1]) have tried to explain the occurrence of the macula coloboma as

[1] "Ueber ein Colobom der inneren Augenhäute ohne Colob. d. Iris." Dissert, Frankf., 1871.

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due to a rotation of the eyeball, and a consequent displacement of the fœtal cleft; but I am convinced that when I have mentioned some of the cases which have come to my notice, it will be clear that this theory must be abandoned. Hoffman has floated the ingenious theory that colobomata confined to the choroid may be due to intraocular pressure, tearing asunder the barely closed fœtal cleft; but here again, apart from *a priori* objections, many of my cases are incompatible with this hypothesis. Schweigger's theory,[1] which might be termed the "fœtal inflammation" theory, is very widely accepted at the present time, and overcomes many difficulties in the way of the other suppositions, and yet, although affording a probable clue to certain cases, will, I believe, fail in accounting for some of the cases I hope to show.

The first point which I think I can demonstrate is that extra-papillary colobomata can occur in any part of the fundus, and I believe that the number of cases which have already come to my notice are sufficient to authorize my making such statements.[2] Besides the colored drawings of twelve cases, published with this paper, I have in my possession a number of other drawings similar in character to these.

The two following diagrams represent the fundus of the right and left eye, and I have indicated thereon a number of the extra-papillary colobomata which I have seen, as well as some few which have been illustrated by previous writers. I may, however, add that those illustrated by previous writers are, with scarcely an exception, found in the region of the macula.

All the above cases, with five exceptions, have come under my own observation. Two of these exceptions, Nos. 4 and 20, are inserted by kind permission of Prof. Tweedy, and the three remaining are copied from the drawings of

[1] Schweigger: "Lehrb. d. Augenheilkunde."

[2] The same remarks apply equally well to iris-colobomata. The usual direction we know is downwards, and deviations from that direction were considered not to be colobomata (Manz: *Graefe-Saemisch. Hand-book*, II., 2, p. 65); but v. Mittelstädt has described cases occurring both outwards and inwards; and Prof. Tweedy assures me he has met with an undoubted congenital iris coloboma directed vertically upwards.

Diagrams showing the relative size and position of various colobomata that have come under my notice or have been elsewhere figured and described. To avoid confusion the very numerous "central" colobomata have, with two exceptions, been omitted. The ordinary "fissure' colobomata have also been left out.

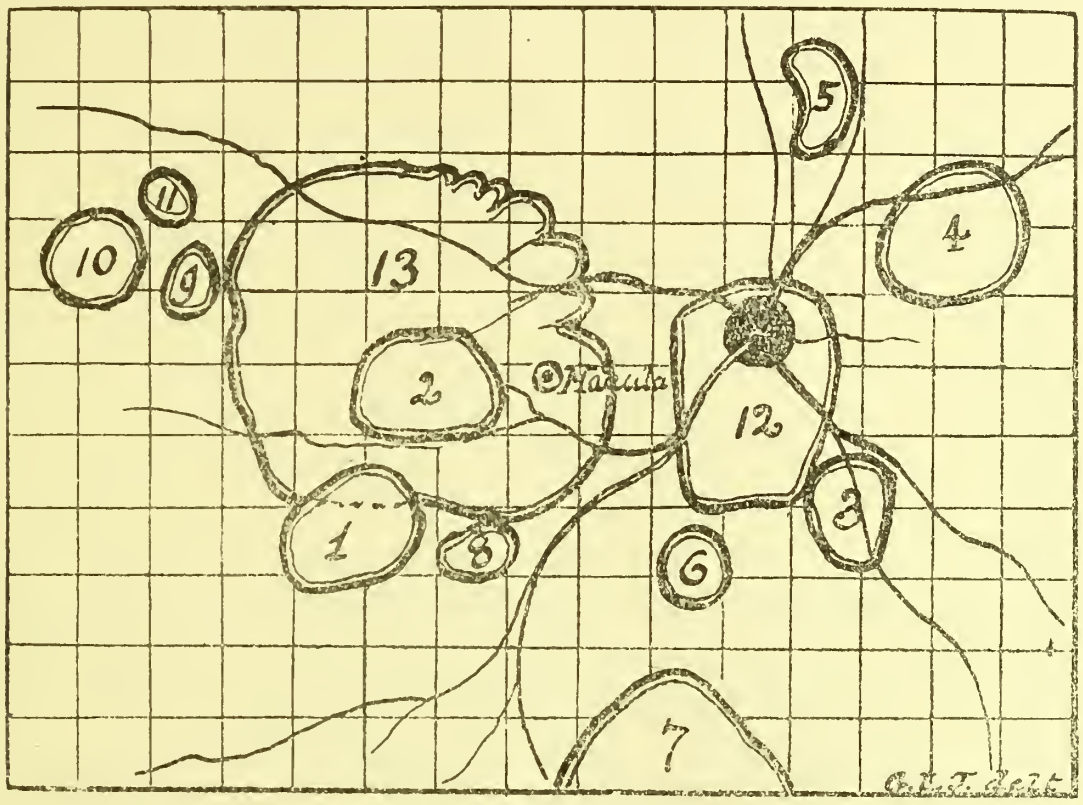


FIG. 1. R. EYE.

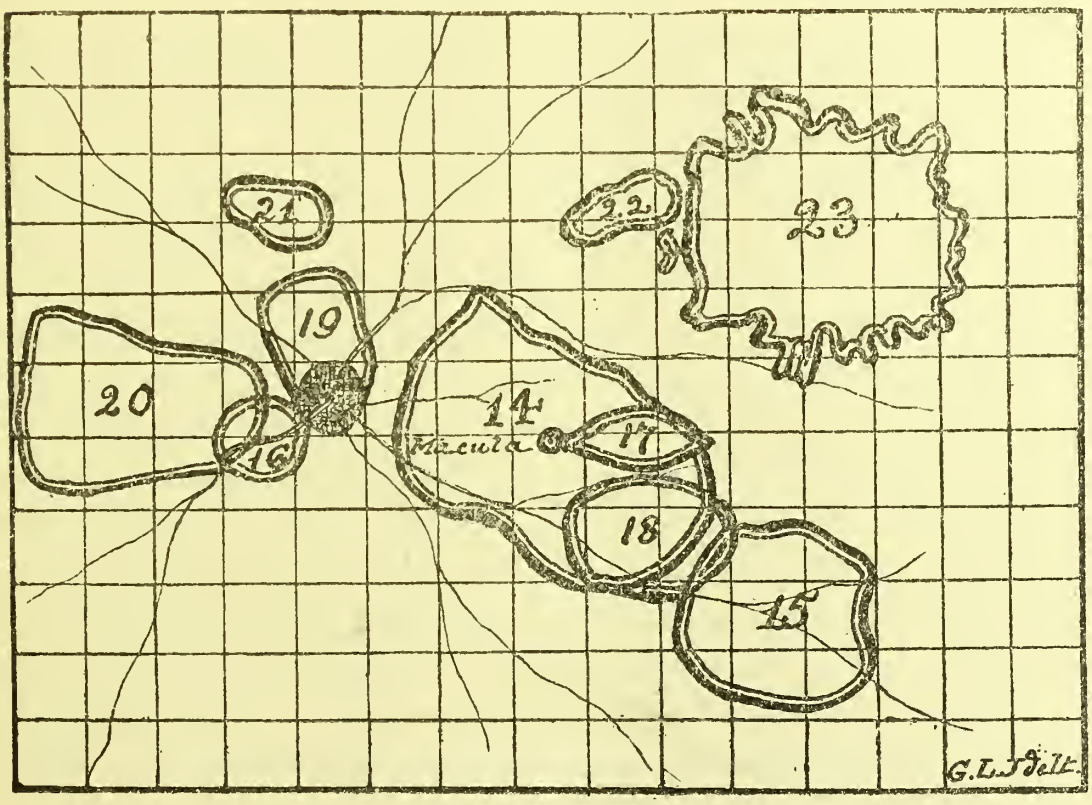


FIG. 2. L. EYE.

EXPLANATION OF DIAGRAMS.—FIG. 1.—R. EYE (erect image). 1. Pl. I., Fig. 1. 2. Pl. IV., 7. 3. Pl. V., 10. 4. Mr. Tweedy's 1st case. 5. John B——. An unpublished case drawn and described by me at Moorfields. 6, 7. Hirschberg's case figured in the *Centralbl. f. p. Augenh.*, Aug., 1885. 8, 9, 10, 11. The remarkable case of W—— S——. Pl. VII., 13. 12. Papillary coloboma, Pl. VI., 12. 13. Macula coloboma. Pl. VII., 13. FIG. 2.—L. EYE (erect image). 14. Macula coloboma. Pl. VIII., 14. 15. Extra-papillary coloboma. Pl. VIII., 14. 16. Schweinitz and Randall's case. A. OF O., Vol. xvii., Part 4. 17. Pl. V., 9. 18. Pl. II., 4. 19. Pl. IV., 8. 20. Mr. Tweedy's 2d case. 21, 22. A recent case. A large very characteristic "macula" coloboma was present in the same eye, and a papillary coloboma in the right eye. Unpublished. Found and described by me at Moorfields. 23. Fuchs' case. See ARCHIVES OF OPH., 1883, p. 37, with plate.

Prof. Hirschberg, Prof. Fuchs, and Dr. Randall. Only two central colobomata have been drawn (Nos. 13 and 14), which, being larger than any previously described, would probably include in their circumference all the rest. The typical "fissure" colobomata have likewise been omitted. The outlines of the colobomata are drawn accurately to scale, and to assist the reader in grasping their size the diagrams are divided into squares, each the size of the average optic disc. Some of the colobomata are very large. One has only to compare the small circle (No. 13)—de Schweinitz and Randall's case—with their original drawing in the ARCHIVES, to appreciate the distance from the macula or disc at which congenital gaps may be found. I have to thank Dr. Paul J. Sartain, of Philadelphia, for first calling my attention to the case of Sarah S——, who came to the Western General Dispensary complaining of blepharitis. Suspecting hyperopia he examined her refraction with the ophthalmoscope, and this led to the discovery of the coloboma.

Prof. Tweedy's first case was discovered in a still more curious way. A man fell off the raised platform of one of our railway stations in attempting to turn round as he was walking with the railway line, on his left hand, and on recovering from the shock, thought he had injured his right eye. This resulted in an action for damages against the railway company, the plea being that the imperfect manner in which the station was lighted had contributed to his fall. and set up "choroiditis" in the eye. In the evidence, Mr. Tweedy stated that so far from the accident having caused the patch of "choroiditis," the so-called choroiditis had been the cause of the accident, it having evidently existed from birth, and, being situated upwards and inwards to the disc, had, owing to the scotoma it produced, prevented his seeing the edge of the platform, which was below and to his outer side. This will, I think, show that a careful study of these defects may have a practical issue.

Mr. Tweedy's second case, that of Rudolph D., æt, twenty, was examined ten years ago (1879), and then pronounced by him (in direct opposition to the opinion of

a very eminent London oculist, who maintained that "the back of the eye was undergoing degeneration") to be a congenital malformation. Mr. Tweedy's last notes of the case two years ago show that no change whatever has taken place, the vision with + 1 D still being = $\frac{6}{6}$. In this case the remnants of a hyaloid artery, in the form of a white fluffy cord, could be seen stretching from the disc to the posterior capsule. A coloboma of the optic-nerve sheath was also present in this eye; the other eye was slightly myopic, R V with — 1 D = $\frac{6}{6}$, and showed no traces of any other congenital defect, except that the outer third of the disc was pigmented.

Nos. 21, 22 occurred in a girl, æt. twenty-two, who came to Moorfields for spectacles, as a patient of Mr. Lawson. Not getting more than $\frac{6}{18}$ with any glass, I examined the fundus, and discovered three characteristic punched out colobomata, one 3 P D's oval horizontally at the macula, one upwards and 3 P D's to the inner side of the disc, and one 3 P D's upwards and 3 P D's to the outer side of the disc. Except for a myopic crescent, and some shrinking of tissues on the outer side of the disc, there were no other defects.

I take it for granted that the reader will admit, in looking at the above diagrams, that there is reason for concluding that colobomata may occur in *any* part of the fundus, although the favorite seats are undoubtedly the region of the macula lutea and the choroidal fissure.

Reasons for the Macula Being a Favorite Seat.—Although one would naturally expect to find a coloboma remaining to bear witness to an imperfectly closed choroidal fissure, the reason why the macula should be so frequently the the seat is at first sight more difficult to account for; since von Ammon's rotation theory, as we have above said, only creates fresh difficulties. Many ophthalmologists, recognizing these objections, have endeavored to explain the macula colobomata on the hypothesis of intra-uterine choroiditis.

I think it unnecessary to go out of our way to seek the explanation in a disease which after all is only a presumption, and about which we know next to nothing, when we can find some facts to assist us close at hand. In the first

place, the region of the macula is by nature a weak one. Hannover, in his "Memoir on the Funiculus Scleroticæ," says: "I have come to the conclusion that the fovea (centralis) is not only a formation due to an arrest of development, but also the most defective part of the entire retina (det mest defekte sted i hele Nethinden)."[1] According to him, this funnel-shaped cicatrix is situated in the sclerotic, immediately behind the fovea centralis, passing through the entire thickness of the sclerotic, and ending freely outside the globe in a kind of tag or short cord. He says he has examined 50–60 eyes, and has never found it absent. "One can also find traces of the fœtal cleft in the other portions of the sclerotic in the neighborhood of the funiculus . . . and these characters are still more pronounced in the eyes of the newly-born." He adds: "In the coloboma of the eye I have shown that the deep oval pit seen in the inside of the sclerotic behind the fovea, and the thinness of the membrane at this point, are due to an arrest of the (fœtal) cleft of the eye." It is not difficult to believe that in the formation of the secondary optic vesicle, this weak spot at the macula would be more likely to give way than any other part of the retina. Van Duyse even goes so far as to place the macula and the optic nerve in the middle of the embryonic fissure, and believes that in a normal eye the fissure to the outside of the optic nerve gets separated off from the embryonic fissure by the interposition of retinal elements.[2] I think this supposition lacks confirmation, and is certainly difficult to reconcile with Würzburg's interesting discovery of an embryonic macula lutea on the inner as well as the outer side of the papilla,[3] and still more so with the changes which these symmetrically placed yellow spots subsequently undergo.

In the second place, traces of arrested development apart from the coloboma may be seen in the great majority of cases. Thus, out of sixteen consecutive cases which I

[1] Funiculus Scleroticæ en Levning af den fœtale Spalte i Menneskets Oie. Efterviist af Adolph Hannover. Meddelt i Videnskabernes Selskabs Mode d. 8. Dec., 1876.

[2] Du Colobome Central ; *Ann. d'Oculistique*, Jan., 1884, p. 32.

[3] On the Development of the Eye in Mammals. A. OF O., 1887, p. 230.

have met with, three of them showed traces of a hyaloid artery (see Plate III., Fig. 5); in four cases the eye was distinctly smaller than its fellow (see Plate II., Fig. 3; Plate VI., Fig. 11). In considerably more than half the cases the retina and choroid appeared thinned, or atrophied, round the margin of the disc, sometimes amounting to a distinct coloboma of the nerve sheath. Plate I., Fig. 2; Plate II., Fig. 4; Plate V., Fig. 9; Plate VII. and Plate VIII. The same may be observed among the cases described by Streatfeild, Randall, Montmeja, de Wecker, several of Van Duyse's Fuchs, Jessop, Tweedy, Hirschberg (1st case), Schnabel, Reich, and many others. Moreover, I noticed opaque, or at least translucent, nerve fibres in four cases. (Plate III., Figs. 5 and 6; Plate IV., Fig. 7.) See also van Duyse's case, *Annal. d'Ocul.*, No. 94. And in two cases (Plates VII. and VIII.) the whole outer portion of the disc included between the art. temp. sup. and art. temp. inf. was occupied by a network of choroidal veins, while the retinal pigment was more or less deficient right up to the coloboma.

It is curious that in no case of extra-papillary coloboma have I noticed any deficiency or change in the iris. Remak accounts for this fact on Hoffman's supposition that the intra-ocular pressure had torn asunder the barely closed fœtal cleft.[1]

Refraction.—Most of the cases were hypermetropic. Out of my sixteen consecutive cases three were emmetropic, three myopic (viz, 12 D, — 7 D, and — 9 D), eight hypermetropic, and two had mixed astigmatism. Now a small or undeveloped eye is usually associated with hypermetropia, and, consequently, this is what one would expect in colobomata of *small dimensions*. On the other hand, in *extensive* colobomata the coats of the eyeball would be seriously weakened, and tend to stretch and become staphylomatous, and thus directly induce a myopia, and that is what I found to be the case.[2] See Plates VII. and VIII. and Plate I., Fig. 2.

[1] Ein Fall v. Colob. der Mac. lutea; *Centrabl.*, Sept., 1884.

[2] I think it must have been this latter class of cases which Schnabel had in his mind when he traced the connection between the etiology of myopia and the so-called coloboma circa maculam. See Schmidt-Rimpler, *Graefe's Archiv*, vol. xxvi., 2, p. 221.

And moreover the degree of myopia would be in proportion to the proximity of the coloboma to the posterior pole of the eye. Hence the myopia is considerable in the cases shown in Plate VII. and Plate VIII., while absent or nearly so in Fuchs' case, (see diagram L. E., No. 23). On the other hand, if the coloboma is of moderate size, *i. e.*, 1 to 3 P D's only, it may not affect the refraction at all.

Field of Vision.—The importance of ascertaining the exact sensibility to light over the coloboma area is obvious. Schmidt-Rimpler points out that it is not sufficient in order to establish a coloboma that one should have found a scotoma by means of the ball (or flag), but that every sensation of light, whether from the lamp or ophthalmoscope mirror, must be absent.[1]

Finding the signal made of white paper, commonly used, reflected far too feeble a light to be of decisive value in such an experiment, I constructed a small electric lamp for the purpose. See sketch opposite.

If these eight charts (which are samples of the rest) be examined we shall notice several facts:

1. The electric-light area for the whole field is slightly larger than the area for the white flag (reflected light) (see Charts 1, 3, 4, 6, etc.).

2. The electric-light area of the scotoma, though in every case smaller than with the flag test, is nevertheless quite definite, and shows that although the marginal retina (the pigment area) round the coloboma is sensible to the incandescent light, it is insensible to the stimulus of the reflection from the white flag (see Charts 1, 3, and 4).

3. Inside this zone the field is insensible to the most powerful stimulus.

4. In many cases the V F is very limited, and does not include the scotoma in its area, and yet a well-marked notch in the boundary line exists, both for the lamp and the flag in the region of the field corresponding to the coloboma, but somewhat nearer the axis of vision (Charts 1 and 2).

5. In one case no scotoma for the flag existed, because

[1] *Augenheilkunde*, 3te Auflage, 1888, p. 236.

the section of the field in which the scotoma should lie was contracted within its inner boundary, and yet by the enlarged field given by the electric light the scotoma could be clearly brought out (see Chart No. 6). Compare also Chart No. 5, where scotoma is unseen for red and green, but brought out by white flag.

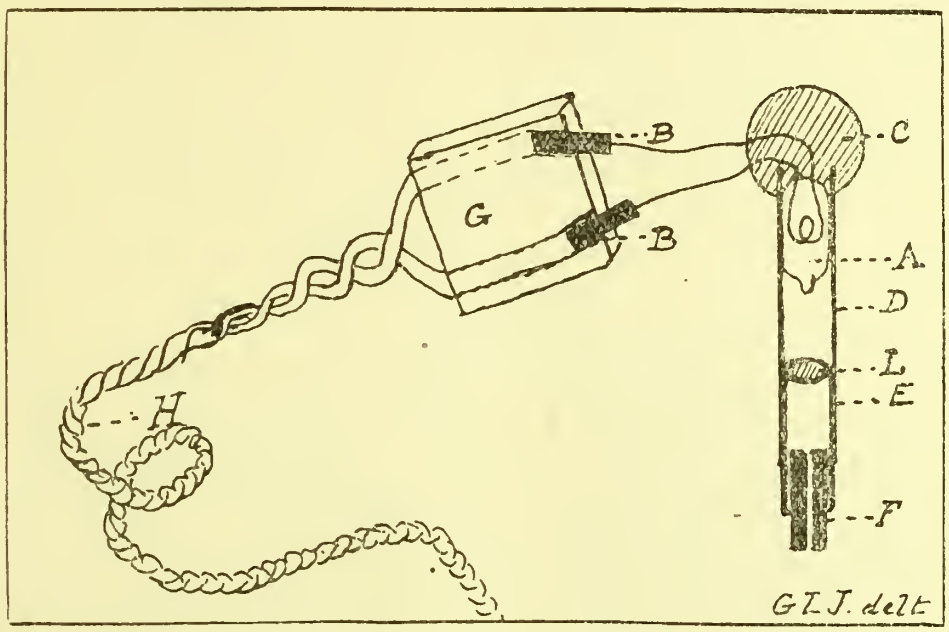


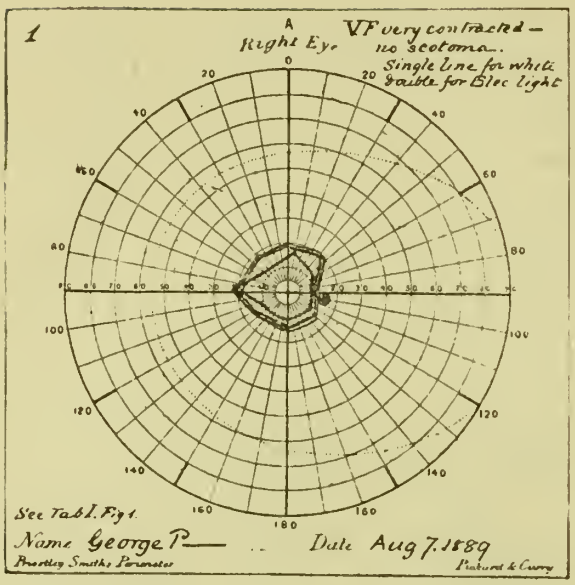
FIG. 3.

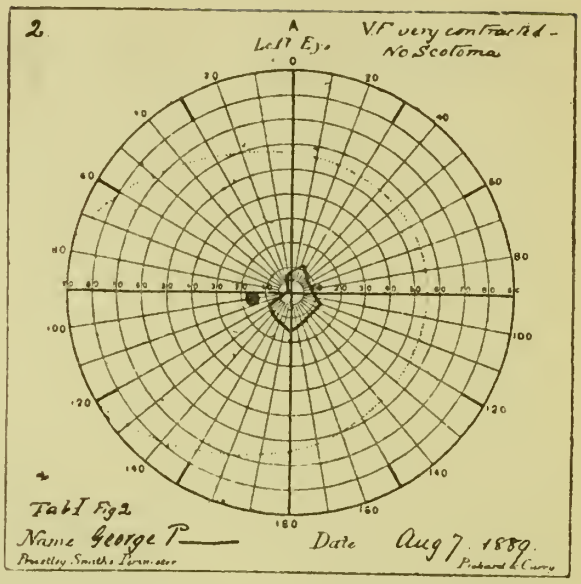
Fig. 3. Incandescent condensing lamp, used for ascertaining the area of partial and total insensibility to light in colobomata.

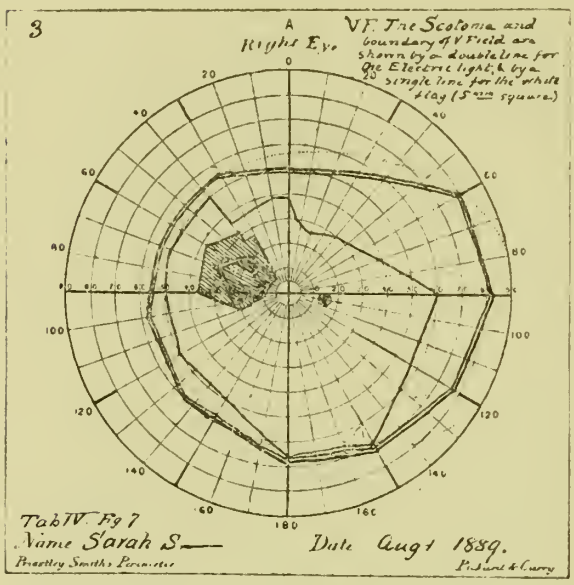
A is a very small incandescent lamp, which is fixed in position at the end of the brass tube D by means of a lump of plaster-of-Paris. This lamp is connected with the double wire H from the battery by means of two small wires which end in the short rods B B, fitting the two brass tubes in the block of vulcanite G. A second tube E slides inside the tube D, and at its end is fitted a biconvex lens of $\frac{3}{4}$ inch focus. By sliding this inner tube the lens may be placed exactly at its solar focal length away from the light A, and thus render all the emergent rays parallel. In order to limit the area of light still more, I drilled a 1 *mm* hole lengthways through a short rod of brass F, which fitted the second tube, and blacked the inside. To use the lamp the tube D slides into a fourth tube, which is soldered on to the runner which slides along the arm of the perimeter. This soldered tube was split along its length to admit the lamp tube easily, and was carefully directed, so that in every position along the arm the tube is in a line with the pupil of the patient's eye. The lamp is of two-candle power, and is worked by a storage battery of four volts. I have had it in use for several months and find it works admirably.

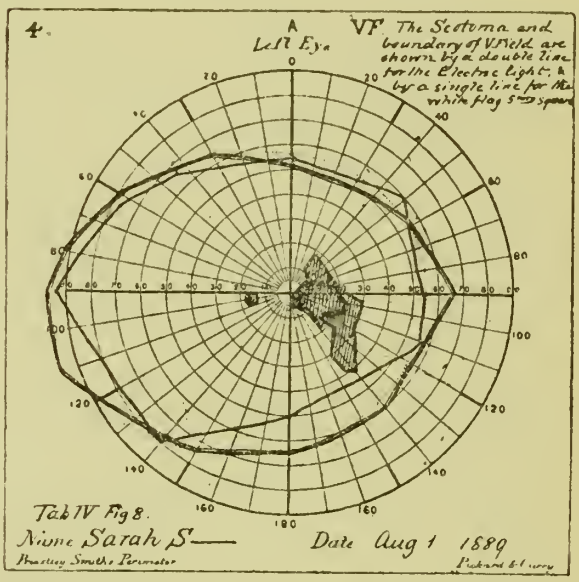
Lastly, very few of the scotomata corresponded in area or shape with the colobomata, as seen with the ophthalmoscope. This, I admit, is directly contrary to the observations of Schmidt-Rimpler,[1] De Wecker,[2] and others, but I made

[1] *Graefe's Archiv*, xxiii., 4, p. 176.
[2] De Wecker et Landolt: "Traité Complet d'Oph.," vol. ii., p. 515.





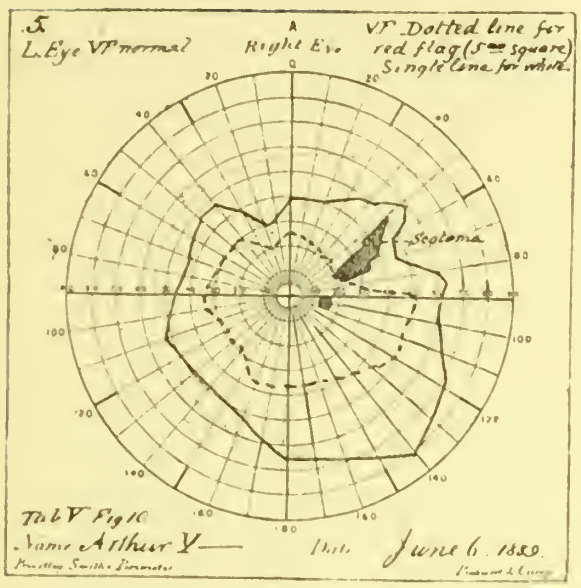


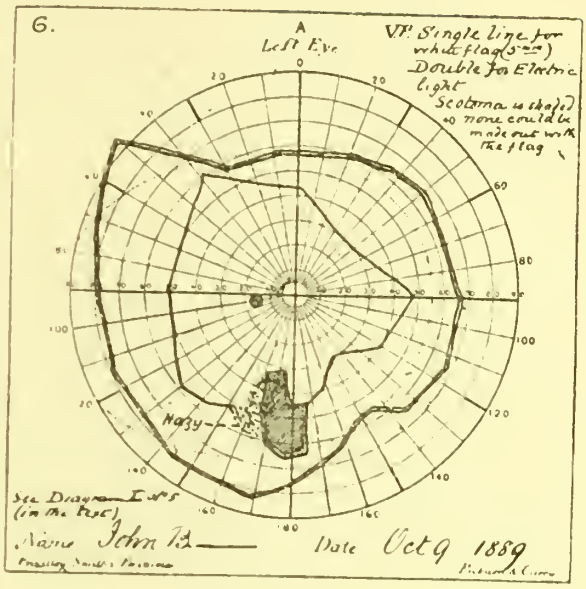


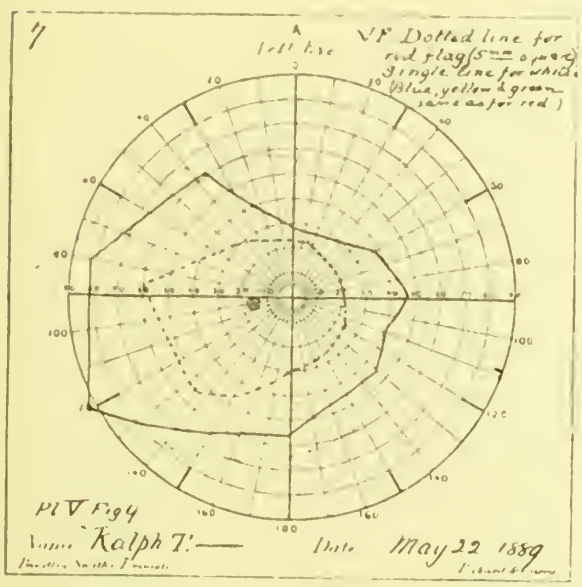
my observations so carefully that I have no doubt as to the correctness of the results.

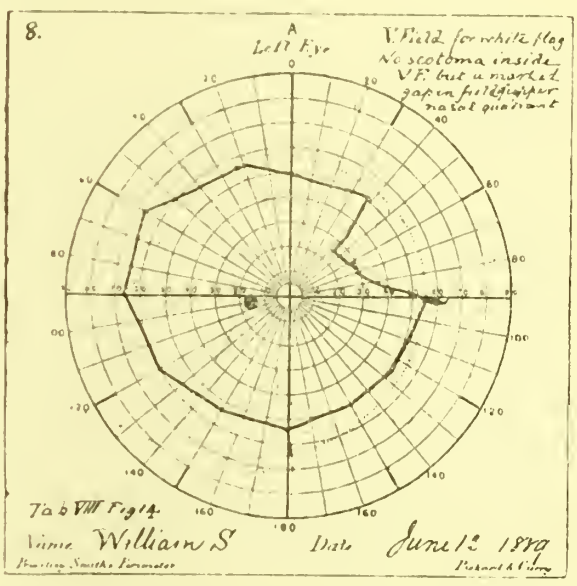
Charts 3, 4, and 6 are the only ones out of over thirty in which the scotomata even approximately resemble the shape of the coloboma, and that only held good for the electric light; white and red, green and yellow, giving shapes not only several times larger, but bearing no exact relation in position or shape to the coloboma as seen with the ope. directly.

Diagnosis.—It now becomes necessary to determine in what manner extra-papillary colobomata can be diagnosed so as not to be mistaken for cases of choroiditis and other inflammatory conditions of former and recent dates. Now,









the only diseases which could possibly be mistaken for as coloboma are:

A. Sclero-choroiditis posterior.

B. The plastic (exudative) group of choroidal affections, especially in their later (atrophic) stages. Under this head we include the disseminate varieties, both simple and specific, central, senile, and areolar choroiditis.

C. Subretinal and choroidal hemorrhages in their later stages.

It is beyond the scope of this paper to describe the above diseases, but I wish to point out the most salient characteristics which can guide us in diagnosing extra-papillary colobomata.

1. The margin of an extra-papillary coloboma is always sharply defined, and it is everywhere surrounded by healthy tissue, while it stands to reason that in disease this can rarely be the case.

2. In all cases of extra-papillary coloboma the pigment is found in front of the retinal vessels, and never behind, whilst in the above-mentioned diseases the pigment is not necessarily confined to any layer.

3. In choroiditis the tendency is towards the formation of multiple lesions, except in those forms such as senile and central choroiditis, which, however, are easily distinguished by other characters.

4. During the active stage of disease the appearance of the lesion naturally alters, whilst a coloboma always remains the same.

5. The most important characteristic of all, however, is the appearance and position of what I may be allowed to term the floor of the lesion. In coloboma one sees the sclerotic of a dazzling whiteness, forming the floor of a well-defined punched out pit, sometimes covered with a layer of connective tissue in a mother-of-pearl-like circular patch. In most cases of choroiditis the floor is of a dirty yellow color, or if chalky white other characteristics, such as the plurality of the lesions, and especially their small size will prevent any mistake.

Moreover, in choroiditis the depression, if any, is much less marked, the lesion often fading insensibly into the surrounding tissue, and often without any marginal layer of pigment. It has been pointed out, I think by Schnabel, that the margin of a coloboma is bounded by a whitish zone of atrophied tissue, and when seen is quite characteristic. I have certainly noticed it in some of my cases, but by no means in all.

I must here repeat that I am only dealing with extra-papillary colobomata, being aware how difficult it is to distinguish many colobomata of the optic nerve sheath from myopic crescents in connection with Sclero-choroiditis posterior.

In connection with the position of the floor, I would re-

mark that, in *all* those cases in which the vision was normal and in some of the cases where it was deficient, I have found the difference of the focus between the margin of the coloboma and the floor amounted to between 2 D and 3 D.

By Dr. Landolt's tables for axial ametropia, based on the well-known formula $\vee - \varphi = \frac{\phi\,\phi'}{\mu - \phi}$, we find, by placing a lens of — 1 D at the anterior principal focus of an emmetropic eye, that the image is removed to a point 0.321 *mm* behind the retina. From the table of eye measurements in Quain's Anatomy we find the thickness of the sclera, choroid, and retina together = 0.08 in., and of the sclerotic alone = 0.05 in., giving for choroid and retina together a thickness of 0.03 in. = 0.75 *mm*. Now, if 0.321 *mm* corresponds to a correcting lens of — 1 D, a plane removed 0.75 *mm* farther back will need a concave lens of 2.5 D placed at the anterior focus.[1] This closely coincides with the actual lens used in the ophthalmoscope to bring the floor of the coloboma into focus. Of course, these figures are only to be considered approximately true, as we find it impossible to focus the fundus accurately within half a dioptry, and, moreover, it is open to doubt whether the thickness of the retina ought to be taken into account at all; but, making every allowance, I think it shows that in many cases of coloboma the depression was not an ectasia or staphyloma, but was *due to the absence of retina and choroid*, enabling us to see the sclerotic in its unaltered position.

Vision.—If the reader takes the trouble to glance over the notes of the cases appended he cannot but be struck with the excellence of the vision in many of the cases. Thus, in the case of Sarah S—— (Figs. 7 and 8, Plate IV.) and of Ralph T—— (Fig. 10, Plate V.) the vision was above normal = $\frac{6}{5}$ in both eyes, while in that of George P—— (Fig. 1) and George G—— (Fig. 3), as well as in Professor Tweedy's two cases (see diagrams), the vision = $\frac{6}{6}$. In Arthur Y——'s case the vision was nearly $\frac{6}{6}$. This was

[1] The anterior principal focus is situated 13.7 *mm* in front of the cornea, which is as nearly as possible the position of the correcting lens of the ophthalmoscope when used in the direct examination of the fundus.

remarkable, as the coloboma encroached on the fovea itself, while in the case of Sarah S—— the coloboma were very near the fovea. I think such cases would argue strongly in favor of a congenital defect, as I have never known of a case of choroiditis close to the macula in which the vision was at or above normal.[1] I have not noticed any alteration in vision unless the coloboma actually covered the macula region.

I might supplement my remarks under this heading of diagnosis by a reminder that, although we must not expect to find any pathological changes in a coloboma, yet the mere fact of the presence of a defect is sufficient to render the particular portion of the fundus more liable to be attacked by disease. To illustrate this I have inserted two drawings (Plate VII., 5 and 6), in which pathological changes in the neighborhood of the colobomata are present.

Pigment and Nævi.—We find in extra-papillary colobomata, as in many other cases in which the retina and choroid are atrophied, an agglomeration of pigment round the margin, and this pigment, moreover, is frequently distributed over the coloboma in the form of a veil or network which lies above the retinal vessels. In some cases the pigment merely forms a single ring bordering the coloboma; in others it forms a double contour or crescent, separated by an intermediate zone of thinned or atrophied choroid (see Figs. 1, 4, 6, 7, 8, 14). In others, again, the marginal pigment passes in the form of trabeculæ towards the centre of the coloboma, where it forms a fine veil or network. In many cases these trabeculæ are wanting, but their course can still be indicated by a few short processes or threads, as if the trabeculæ had been broken off and absorbed (see Figs. 9 and 14). These remains can be detected in nearly all cases in which the central network is absent, and I have seen them in many of the drawings made by previous writers, although their import has, to my knowledge, never been pointed out.

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[1] Mr. Nettleship, however, describes and figures a case of severe central choroiditis with a vision of $\frac{20}{20}$ nearly. But he adds: "Such changes involving, as they appear to do in this case, the fovea centralis, would be incompatible with such excellent vision as this eye possesses."—Trans. Oph. Soc., vol. v., p. 147.

Under this layer of pigment portions of the choroid are seen in an altered form. The choroidal vessels which pass into view from beneath the free edge of the coloboma unite together to form a dense impenetrable plexus towards the centre of the coloboma, which bears the closest resemblance to nævi seen elsewhere (see Plate I., Fig. 2, Plate V., Fig. 10, Plate VII. and Plate VIII). Nævi of the choroid and other tunics of the eye are not altogether unknown. Thus Lawford[1] gives a drawing of a microscopic section of the choroid in which there are unmistakable signs of a nævus, and he concludes that, "*a priori*, one feels inclined to say that the choroid, with its unusually vascular structure, would be a not unlikely situation for the development of a nævus." Horrocks[2] describes a case in which a port-wine colored nævus in the face was observed along with a nævus in the sclerotic and cornea, while the retinal vessels are extremely tortuous. Dr. Sturge[3] describes a case in which a congenital port-wine mark on one side of the face coincided with tortuous retinal vessels, and changes in the choroid on the same side. Lastly, Dr. Milles[4] mentions a case of nævus of the choroid accompanied by a detachment of the retina. The face was also marked by a large nævus.

Conclusion.—The cases published herewith will, I believe, already suffice to demonstrate that extra-papillary colobomata are not of as rare an occurrence as have been hitherto supposed, and I may add that whilst writing this paper I have observed several other cases, which only confirm what I have herein stated. I believe it will be found that they are not difficult to diagnose, and that with the exception of those cases which include the macula they do not impair the vision beyond causing a limitation of the field (unless they are so large as to weaken the resistance of the globe).

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[1] Trans. Oph. Soc., vol. v., p. 138.

[2] Trans. Oph. Soc., vol. iii., p. 106. See also Perrin and Poncet's "Atlas of Path. Anat. of the Eye," where a case described by them as "Congestion of Choroid" is believed by Dr. Lawford to have been a nævus.

[3] Clin. Soc. Trans., vol. xii., 1879, p. 162.

[4] Trans. Oph. Soc., vol. iv., p. 168.

If I may suggest some explanation of this evidently congenital defect, I think, as I have above indicated, that we are in the presence of something which presents many points in common with cutaneous nævi.

First.—The eye, although a highly differentiated nerve end-organ, is nevertheless essentially a skin formation, while the choroid presents an analogy to the corium both from its position, from its mesoblastic development, and from its affording nourishment to a directly superimposed layer of epiblastic pigment cells.

Secondly.—Wherever an agglomeration of choroidal vessels exists, we find it below the retinal pigment layer, which forms a network over it.

Thirdly.—The vessels forming the agglomerations are so fused together as to be indistinguishable and completely hide the sclerotic beneath.

Fourthly.—The absence of this dense structure in some of the cases can be explained by the analogous occurrence of the altered condition of the skin where nævi have disappeared and become absorbed, and I think it is not altogether unreasonable to suppose that the mother-of-pearl-like sheen, and peculiar glistening tissue which may be seen spreading over the base of some of the colobomata where this vascular mass is absent, is merely the connective-tissue cicatrix of an atrophied nævus.

Fifthly.—Cutaneous nævi are universally admitted to be congenital formations, and where they occur we find an increase of the pigment cells over them. Colobomata are likewise recognized as congenital formations, and the extra-papillary forms, at least, are conspicuous by the pigment clusters which surround and cover them.

Even had ocular nævi never been recorded, I hardly think it would be possible to come to any other conclusion, after examining the cases which form the subject of this paper, than that the peculiar choroidal structure in the centre of certain of these colobomata is of the nature of a nævus.

Lastly, it seems difficult to explain most of these malformations in any other manner, as *in no case could any history be obtained, either directly or indirectly, of syphilis; nor could*

I in any case (except perhaps that of George P——, Plate I.) *obtain the slightest evidence that any inflammation had occurred inside the womb during pregnancy, or that the mother had had any fever whatever.*

Moreover, the lesions have all the appearance of having a congenital origin, and, occurring as they do in every part of the field, cannot possibly arise from any fœtal cleft; besides, the well-known papillary colobomata, whether limited to the sheath of the optic nerve or passing down toward the iris, differ as a rule very materially in structure from the extra-papillary colobomata which form the subject of this paper.

Case 1.—George P., private patient, schoolboy, aged eight, youngest of four children, all healthy. Patient somewhat stunted in growth; head slightly hydrocephalic. Teeth fairly developed, enamel good, no notches. Hair light flaxen, approaching white. Irides blue. Excitable, delicate, subject to headaches. No history of syphilis in parents, nor any signs of it in patient. Right eye well developed; iris normal, active; lens normal. R V = $\frac{6}{6}$ nearly and J 1 at 12 in. No hypermetropia. Ophthalmoscope (see Fig. 1, Plate I.): fundus healthy; choroidal vessels invisible; disc pink, oval; retinal veins full, tortuous near the disc; arteries slightly tortuous in places; Y S small and granular, with well-marked "comet flare-spot."

Six P D's to the outer side, and 3 P D's below the papilla, is a large chalk-white gap, $2\frac{3}{4} \times 2$ P D's, surrounded by a pigmentary zone. This patch is exceedingly white, with a glistening mother-of-pearl sheen; over its surface a faint grayish sprinkling of pigment may be seen, but it is otherwise destitute of pigment or blood-vessels. Three small arteries and several veins can be traced to the patch, on reaching the edge of which they curl round and disappear, as in a glaucomatous disc, but, unlike the latter, they do not appear again. The edge of the white patch is abrupt and very sharply defined, and directly continuous with the surrounding retina and choroid of the fundus. The patch itself is distinctly on a lower level, which may be seen by the obvious parallax, and also by the difference of refraction, — 2 D, or 2.5 D by the direct examination. At the margin the retina advances beyond the choroid, in the form of a crescent-shaped lip, $\frac{1}{8}$ to $\frac{1}{12}$ P D in breadth, and is loaded with black-colored irregularly disposed pigment. The above-mentioned retinal vessels curl over the outer

and thicker lip, and, apparently piercing the deep layers of the retina, run backwards between it and the choroid (*vide* Fig. 1, Plate I.). V F contracted to 20° nasal side, 15° temporal, 20° above, and 10° below; same for electric light, white flag, and colors.

Case 2.—Left eye of above patient, V = $\frac{6}{36}$, J 15 at 6″; sees slightly better at a distance with + 1.25 D. Sight has been defective from birth. The globe is slightly smaller than the right eye.

Ophthalmoscopic appearance (Fig. 2, Plate I.): The disc is slightly smaller than its fellow, round-oval, margin irregular, with faint pigmentary border. The triangle between the A. T. sup. and A. T. inf. is decidedly pale. The outer edge of the disc is bordered by a crescent, which fades imperceptibly into the thinned choroid. This latter forms a considerable patch, extending to at least half-way between the disc and the coloboma, about to be described. Owing to the more or less complete absence of the retinal pigment layer over this patch, the choroidal vessels are very conspicuous. The rest of the fundus (if we omit the coloboma) is quite normal, nor are the choroidal vessels visible anywhere else. At 2½ P D's from the edge of the disc is a large transversely oval-shaped patch, 4 P D's × 3 P D's. This presents some very characteristic points. The sharp bright-red margin of retina and choroid, which can be traced all round the coloboma, is covered with a lacework of dense black pigment, which forms an overlapping fringe. This border is irregular, forming ten well-defined notches filled up by black pigment, and from which as many thick processes or trabeculæ pass towards the central red mass. This latter presents the appearance of a nævus in structure, being evidently largely if not entirely formed of a dense anastomosis of choroidal veins and capillaries. These, for the most part, enter the coloboma beneath the pigment trabeculæ, and, following their course, form the above-described tangled mass. So dense is it that no trace of sclerotic can be seen between the meshes. The pigmentary trabeculæ also join to form an irregular lacework covering the nævus, which can be seen through the interstices, about 1 D below. These lobules, which are mapped out by the trabeculæ, and which bear a rough resemblance to the lobules of the cortex in a lymphatic gland, are, but for a stray retinal pigment and a few choroidal vessels, quite empty, and only show the bare glistering sclerotic shining through. The Y S is evidently absent, being displaced by the coloboma.

V F very contracted for electric light, 10° above, 20° below, 8° temp., 15° nasal. For white flag same-sized field, but different-shaped; slight gap outer side; no scotoma. See chart No. 2.

Case 3 (Figs. 3 and 4, Plate II.).—George G——, aged three and one half. Kindly sent by Dr. de Carey. Patient is a bright and intelligent child, born at full time. Mother perfectly healthy, both before and during confinement. Patient is the youngest of four children—all healthy and have good sight. No trace of syphilis in the family.

Right eye, H 1 D; V = $\frac{20}{20}$ probably [owing to his early age (three and one half years), it was very difficult to get him to answer questions, but several letters of the test line were pronounced]. Disc, fundus, and Y S all healthy. Hyperopic reflex and halo round Y S.

Left eye, barely P. L.; globe undeveloped; cornea about $\frac{2}{3}$ diameter of right eye; refraction with ophthalmoscope and retinoscopy about + 1 D; lens well developed; no sign of cataract; iris blue, acts well to light—entire (Figs. 3 and 4, Plate II.) ophthalmoscope disc lost in a white or faintly pink egg-shaped coloboma 3 P D's × 2 P D's, long axis vertical. The four superior vessels terminate abruptly about the middle of the coloboma. The (numerous) lower ones curl over the lower edge of the coloboma, and, with one exception, disappear. At the place where the vessels curl over is a round glistening mother-of-pearl-like depression, nearly the size of a normal disc, at a somewhat lower level than the rest of the coloboma. The upper part of the coloboma is on a level with the general fundus, the choroid and retina of which do not pass quite up to the edge of the coloboma, but are separated everywhere by a white marginal ring destitute of choroid. A little pigment (retinal) is heaped up here and there round the coloboma. At 1½ P D's external to this coloboma is a boat-shaped choroidal gap or coloboma 2½ P D's × 1 P D, long axis horizontal. This is surrounded by a ring of black pigment and below by a second yellow ring of thinned choroid. The margin is strewn for some distance with fine pigment granules as well. Several choroidal vessels cover the gap from above inwards, and pass out of sight beneath the margin of the gap; the floor of the coloboma is a dirty white and is brought into focus with a — 2.5 D lens behind the mirror (direct); the choroid (and retina?) are thinned, and in three places the sclera shines through between the two

colobomata. Below these the fundus is stippled with pigment, but elsewhere the fundus is healthy. The Y S is lost in the coloboma. The drawing was only obtained after a most wearisome number of sittings, but the result is perfectly accurate (Fig. 4, Plate II.). V. F.: the child being so young it was found impossible to take this satisfactorily.

Case 4 (Fig. 5, Plate III.).—Cathleen W——, aged sixteen, of London. I am indebted to Prof. Tweedy and to Dr. T. Phillips, F.R.C.S., Eng., for allowing me to make use of this case. Family healthy ; six brothers and sisters, all healthy and good sight. Patient always had good health ; eyes normal in appearance, pupils active. Patient came to Moorfields complaining of her eyes aching when reading. L E, V = $\frac{6}{6}$, with + 2.25 D = $\frac{6}{5}$ and J 1 at 16 inches ; ophthalmoscope : fundus and disc perfectly normal. R E, V = fingers only ; eye defective ever since she could remember. Refraction shows + 2 D ; ophthalmoscope : disc normal, pink ; vessels normal. Over region of Y S is a round-oval choroidal gap, the same size as the disc, bordered by a thin zone of pigment. The lower two thirds is nearly dense white, apparently of thickened sclerotic tissue. Two small branches of A. T. inf. pass into it and disappear, and a branch of A. T. sup. crosses it. The upper third is stippled with fine pigment. This coloboma connects the ends of a remarkable C-shaped, glistening, white patch. The upper limb appears to be entirely divested of choroid, and its inner part is limited by the sharp edge of the retino-choroid of the fundus. As we pass round the curve of the C-shaped patch this white deposit lies nearly on the surface and becomes irregularly scattered in a flocculent manner, and no free edge of either retina or choroid is visible anywhere. At the head end of the "C" patch are two nævoid-like patches of choroidal (?) vessels covered over with pigment. This pigment is quite superficial and almost obscures the head end of the "C" patch, becoming less and less in quantity towards the tail end, the last third of which is quite destitute of it. This pigment differs from the usual pigment I have observed in these colobomata, resembling that seen in retinitis pigmentosa. The choroid of the fundus enclosed by the "C" is thinned away in places, showing nearly bare sclerotic. The greater part of the C-shaped patch gives the impression of a deposit resembling that seen around the Y. S. in albuminuric retinitis. Between the coloboma and the disc, the field is traversed by innumerable radiating semi-opaque nerve fibres, while scattered here and there

are a number of yellowish-white dots quite superficial. The presence of these nerve fibres gives support to the supposition that the C-shaped patch is partly congenital, but what proportion is due to persistency of medullary substance or embryonic tissue and what to disease it was impossible to determine.

The urine was tested and found free from albumen, and, moreover, the patient was in the best of health.

Case 5 (Fig. 6, Pl. III.).—Eleanor E., aged twenty-three. Patient came to Moorfield's on account of defective sight in the right eye and was kindly handed over to me by Mr. Lawson. Has never noticed any thing wrong with the left eye. Cannot be sure that she has ever seen well with her right eye, but noticed its defect by accidentally covering her left eye a few weeks ago.

V L = $\frac{6}{6}$ and J 1. Fundus perfectly healthy.

V R = P. L. only.

Ophthalmosc.: R E, disc normal. Vessels slightly tortuous, especially the veins. Arising from near the centre of the disc is a short, thick, bladder-like vessel ending in a cul-de-sac, about $\frac{1}{2}$ P D long, as it appears foreshortened by the ope. Its direction is forwards and downwards, and, illuminated by the mirror, casts a distinct shadow on the fundus. It is only attached to the disc, its anterior end being quite free and movable in the vitreous. At its attachment to the disc there is distinct pulsation, and the whole vessel moves with each beat of the artery. I think I can trace its junction with the main trunk of the inf. retinal A^v.

If it is a persistent hyaloid artery (and I cannot account for it otherwise), it will give force to the claim of my placing the peculiar patch about to be described among my collection of colobomata.

One P D to the outer side of the disc is an oval-shaped depression, occupying the greater part of which is a tongue-shaped, nævoid-like growth, looking like an oyster half removed from its shell. The patch, 2 P D's × 1$\frac{1}{2}$ P D's, has its long axis vertical. The inner and outer margins overhanging the patch are white and glistening. The margins are not white from atrophic changes or deposit, as on careful focussing a great number of fine nerve-fibres are traced across them which can, moreover, be traced over the fundus for a little way. The floor of the patch around the black mass is slightly behind the edge of the patch about 1 D to 1$\frac{1}{2}$ D's only. It is white and glistening, the greater part covered over with fine gray pigment. Slight parallax between the outer edge and the white floor is noticed. Directed obliquely, downwards

and outwards, is a nævoid-like reddish mass, covered with dense black pigment. In the centre of this patch two spots of white (sclerotic ?) shine through where the pigment mesh is open. A few fine arterial twigs cross the coloboma, but they do not curl over the edge, but pass uninterruptedly across. Below the patch and separated 1 P D from it is a crescentic flocculent deposit, quite superficial. Two P D's below this again, abutting the inferior temporal artery, is a single black spot, $\frac{1}{2}$ or $\frac{1}{3}$ P D in breadth. This spot is quite different from the "craters" so commonly seen in specific choroiditis disseminata, having its pigment almost entirely confined to the centre. (The drawing is here incorrect.) Lastly, this is the only spot to be seen in either fundus, while no history or sign of syphilis can be traced in the family. A small hemorrhage is to be seen 1 P D below and internal to the coloboma.

(Since writing this I have again examined the eye, and find now, Oct. 14, '89, that the superficial white deposit has almost entirely disappeared, while the artery which runs along the upper border of the black spot is obscured for about 2 P D's and then becomes well defined. Lastly, five small recent retinal hemorrhages can be traced to a minute artery at the extreme outer edge of the nævoid-like growth.) It may be possible that the changes seen in this case and the C-shaped crescent in the one previously described (Figs. 5 and 6, Pl. III.) are secondary to the true physiological colobomata, which, being spots of least resistance, have attracted, as it were, the pathological changes to their neighborhood.

Case 6 (Fig. 7, Plate IV.).—Sarah S——, aged twenty-two. Patient came to me at the Western General Dispensary owing to her eyes becoming inflamed after much reading. Has never been ill in her life. Mother in good health during each confinement.

V R = $\frac{6}{6}$, with + 2 D = $\frac{6}{5}$.

V L = $\frac{6}{9}$, with + 2 D = $\frac{6}{6}$.

Ophthalmoscope: R E, disc and vessels normal. Fundus everywhere (excluding coloboma) normal. Y. S. dark red, $\frac{1}{6}$ P D; $3\frac{1}{2}$ P D external to disc, and with upper border on a level with macula, is an egg-shaped coloboma 2 P D's × $1\frac{1}{2}$ P D's, long axis horizontal. The coloboma is bounded by a ring of dense black pigment, less in the upper part than the lower, where it spreads for some distance beyond the coloboma over the field in the form

of a large-meshed network, in the meshes of which the choroid is thinned or nearly absent, showing white patches of clean sclerotic shining through. Between the coloboma and the Y S the field is occupied by a large group of radiating opaque nerve fibres which if traced back would meet in a focus about the centre of the coloboma. The Y S which is quite healthy appears as a cherry-red spot at the lower edge of the bundle of opaque nerve fibres just described, and half a P D internal to the coloboma. The choroid fills up the gap of the coloboma, giving it a reddish-yellow appearance, thinning to white below. It is placed, tested by refraction, 2 D's behind the retinal pigment, covering the free edge of the retino-choroid surrounding the gap. Three large choroidal vessels run horizontally across the gap, between which are minute choroidal capillaries. The choroidal vessels which run across the gap disappear from view at either end. A solitary retinal artery passes over the lower edge of the coloboma, and continues its course along the field. The pigment which covers the lower free edge of the gap is very dense and broad, and, collecting into trabeculæ, encloses a number of small gaps or spaces, which closely resemble the main coloboma except that they are much smaller in size. The upper lip of the big gap consists of a double contour or edge ; the innermost and most projecting part consisting apparently of choroid and posterior layers of the retina, while the part anterior to and just outside the deeper lip seems to consist of retina and retinal pigment only. These lips are on a different level. I think this double lip extends right round, but along its lower half and sides the dense black pigment obscures it.

The space between the two lips is quite white.

V. F. contracted, with marked marginal gaps. Well defined scotoma to electric light. For white and colors somewhat larger field and scotoma. See chart No. 3.

Case 7. (Fig. 8, Plate IV.) Same patient as the last, left eye. Ophthalmoscope : fundus almost identical with that of right eye and (excluding coloboma) everywhere normal. Y S well-defined, reddish spot with bright yellowish glistening centre ; ½ P D below Y S and just external to it is a large egg-shaped coloboma, identical in position and shape with the one in the right eye. It is surrounded by a fringe of dense pigment, which is thin below, but spreads out from its inner margin into an irregular mass occupying nearly the area of the coloboma and extending upwards to

nearly the level of the Y S. This mass contains four oval gaps nearly destitute of choroid, which allows the white glistening sclera to shine through. The lower and the inner of these gaps are almost hidden by dense pigments, but the other two show the white sclerotic base about 2½ D's behind the sharp edge of the retinal pigment. The base of the large coloboma is due to an immense number of fine stippled dots, uniform in size, but which get fainter and fewer towards the centre of the patch. The lower third is crossed horizontally by several horizontal choroidal vessels fringed with thin gray pigment dots. L E, V. F. perfect in extent. Well-marked scotoma to nasal side. See Chart No. 4.

Case 8.—Ralph T——, aged, 19 clerk. Patient came to Moorfields, (to Mr. Lawson's clinic) complaining of inability to see objects at a distance. Strong, healthy, never been ill. Teeth well developed, regular, no absence of enamel. Eldest of five children, all well and good-sighted. No history of mother having been ill before or during pregnancy.

V R with — 5 D S = $\frac{6}{9}$ and $\frac{6}{6}$ nearly.

V L with — 6 D S and — .75 D — C = $\frac{6}{9}$ well and $\frac{6}{6}$ a few letters.

Irides bluish-gray, normal. Pupils active. Both eyes well developed. Ophthalmoscope : R E, fundus shows slight thinning of choroid, and retinal hexagonal pigment round the margin of disc, especially on outer side, where there is a pronounced crescent, otherwise the fundus is quite healthy. No trace of coloboma or other defect anywhere. Y S dark red, slightly granular, with yellowish-white centre spot.

Left eye (Fig. 9, Plate V.).—Ophthalmoscope : disc somewhat smaller than in right eye, and ill defined at margin. Wedge-shaped outer triangle of disc pale bluish-white, bordered by well-marked myopic crescent, with double border edged with pigment. Fundus thinned for a considerable distance all round disc, almost reaching the Y S on the outer side. Y S situated farther away from the disc than usual—viz., 3½ P D's to its outer side. When sharply focussed a number of fine radiating striæ can be seen passing from the fovea in all directions, stippled with minute pigment dots. Commencing from the outer edge of the macula, extending horizontally outwards, is a well-defined gap 2 P D's × 1¼, long axis horizontal. It forms a gap in the fundus like a button-hole stretched open ; the inner angle of which " fades " almost imper-

ceptibly into the Y S, the outer angle being more sharply defined and covered with a black patch of pigment. The curved borders above and below are fringed with black pigment, and sharply defined, and overhanging the floor of the gap. Running horizontally across the floor are three broad anastomosing choroidal veins, which are again crossed at right angles by three fine retinal vessels which can be traced to the A. T. inf. A fine gray pigment mottling can be traced for some distance all round the patch. The whole base of the patch is pale ; the sclera shining through as a faint pink, becoming in places almost pure white, having the appearance of the fundus of an albino.

V F : No scotoma could be made out, although every possible care was taken, and the field tested separately for white, red, blue, yellow, and green. Outer half of field for white nearly normal, inner half contracted to 15°. Field for colors all equal and contracted to 60° out and 20° in. See Chart No. 7.

Case 9 (Fig. 10, Plate V.).—Arthur Y——, warehouseman, aged twenty-five, came to hospital complaining of floating threads (*muscæ volitantes ?*) when looking at the light. Four brothers and two sisters all good sight and healthy. Mother died a few years ago of Bright's disease. Patient always had good health and sight. Teeth fairly well developed, no spaces, or notches. V R = $\frac{6}{5}$ perfectly. V L = $\frac{6}{5}$ perfectly. No manifest hyperopia. Eyes well developed. Irides brown, perfect, act well to light.

Ophthalmoscope : L E, fundus and disc absolutely normal.

Ophthalmoscope : R E, disc round, very pink vessels, tortuous and glistering (shot-silk sheen), otherwise healthy. Y S healthy, surrounded by a pale halo. Fovea yellowish, pin's-point in size. Fundus healthy everywhere except below the disc, where the choroidal vessels show through the retina, in the meshes of which vessels is some very finely stippled *choroidal* pigment. In the centre of this patch of choroidal vessels, 2 P D's below and 1 P D to the inner side of the disc, is an irregular pear-shaped choroidal gap 1 P D vert. × $\frac{2}{3}$ P D horizontal. This gap is surrounded by a sharp margin, over which is distributed a moss-like layer of black pigment ending round the gap. The latter may be divided into two equal parts—an upper and a lower. Upper half shows a white sclerotic base covered with retinal pigment. The lower part of the gap is entirely filled with a mass of choroidal vessels in all respects resembling a nævus. Choroidal veins may be traced to it from all sides round the coloboma.

The retina ends sharply at the margin of the gap, although its pigment may be traced as a thin veil over both upper and lower parts of the coloboma, and is heaped up around it. The thinned choroid extends some little way beyond the scotoma. The rest of the field is quite normal.

L E, V F : Perfectly normal for white and colors.

R E, V F : Irregularly contracted above and to the outside. In the upper and outer segment is a well-marked scotoma for white. The fields for red and green are identical. They are contracted with a well-marked gap where the scotoma should be, but the latter being outside the range for colors, it was impossible to map it out. See Chart No. 5.

Case 10 (Figs. 11 and 12, Pl. VI.).—Flora J——, aged twelve. A private patient kindly sent to me by Dr. de Carey. Patient has always enjoyed good health. Brothers and sisters, eight ; all well. No history of syphilis in the family. L E well developed, iris and fundus normal. R E about two thirds the size of L E. Iris complete, contracts imperfectly to light, enlarges only to 4 *mm* under atropine ; eyelids small, but otherwise normal. Plica semilunaris undeveloped. Slight alternating convergence. V L = $\frac{20}{30}$, V R counts fingers at two feet ; slightly improved with — 10 D. Ophthalmoscope : fundus seen best with — 9 D. Chloroidal vessels can be seen everywhere through the retina. Y. S. not obvious. No halo or fovea visible. Disc injected, ill-defined outline quite invisible below. Is entirely surrounded by a large oval coloboma with flattened ends, surrounded by a border of dense black pigment, which encroaches on the fundus below and at the inner side, but overlaps the coloboma along the outer side. Coloboma measures $3\frac{1}{2}$ P D's × $2\frac{1}{2}$ P D's, long axis vertical, base 2.5 D below surrounding fundus, and chalky white, showing bare sclerotic, except inside lower and outer margins, which are covered with a network of choroidal (?) vessels stippled with pigment granules. The retinal vessels all vanish as they approach the centre of the disc. They are numerous and all spring from various parts of the disc, quite independently of one another, a characteristic feature which is quite peculiar to the papillary form of coloboma, and only after crossing the coloboma do they branch dichotomously in the normal manner. Some very fine pigment-granules may be seen stippled over the field between the Y. S. and the coloboma.

V. F : Vision was too imperfect to get any field.

Case 11 (Fig. 13, Pl. VII.).—William S——, aged fourteen; errand boy. Patient healthy all his life. Teeth quite perfect, regular and even. Eyes normal size. Irides well developed, hazel-brown, pupils contract well to light. Slight nystagmus L E. No history or signs of syphilis in the family.

V R with — 1 D S and + 1 D C axis vertical = $\frac{20}{50}$.
V L with — 1 D S and + 1 D C axis vertical = $\frac{20}{50}$.

R E. Ophthalmoscope: disc round, rosy-pink, vessels normal, well developed. At the outer part of the disc, between the two temporal arteries (A. T^s. and A. T_i.), where they emerge from the centre of the papilla, the latter appears deficient, and in its place we see none of the rosy-pink of the disc, but a dense white base covered with four or five orange-red vessels, which are seen to be continuous choroidal vessels immediately external to the disc, which are plainly seen for some distance (1½ P D's), owing to the thinning of the overlying retinal pigment layer.

At 2 P D's external to the disc is the inner boundary of an immense coloboma measuring 5 P D's each way. This patch fills up nearly the whole space between the superior and inferior temporal arteries. Indeed, the whole character of the fundus is altered between these two arteries, from their commencement to as far outward as they can be traced with the ophthalmoscope. Thus at the centre of the disc we notice at least five choroidal vessels crossing the disc between the two temporal arteries. Tracing the fundus outwards the choroidal vessels are very evident, and the retinal pigment is more or less absent.

Just external to the large coloboma are two more colobomas with a trace of a third. The large coloboma is sharply defined by the edge of choroid, which forms a free edge all round the coloboma. The portion nearest the disc is glistening white and nearly circular, and is situated 3 D's behind the tissue on the margin of the patch. Here the parallax is strongly marked. The patch is bounded externally by a large nævoid mass of choroidal vessels. This structure occupies about half the patch and is covered by a loose veil of retinal pigment. This latter forms the usual ring round the free edge of the coloboma. The upper part of the coloboma is lobulated, and the pigment fills up the recesses between the lobules, and passing between them in the form of trabeculæ, joins the central network of pigment which covers the nævus. Here we may notice the same structure which is so well marked in some of the other cases (see Fig. 2, Pl. I., and Fig.

14, Pl. VIII.), viz., a central nævoid mass covered with a veil of pigment, which latter is connected with the border circle of pigment by trabeculæ; these divide the white zone outside the nævus into numerous lobules as in the cortex of a lymphatic gland. These lobules vary in size and for the most part are oval, with their long axes to the centre. A large number of choroidal vessels may be seen passing from beneath the upper free edge of the patch in a direction parallel to and behind the trabeculæ, to join the meshes which make up the nævus.

Just external to the large coloboma are three small ones. The innermost of these is the best defined. Its base is 2 or $2\frac{1}{2}$ P D's below the surface and crossed by choroidal vessels, and patches of pigment connected with the usual bordering. These three small colobomata differ from most of the others I have examined, inasmuch as the choroidal vessels can be traced up to and beyond the patches, and not merely across them to disappear beneath the margin. This may be explained by the choroid being everywhere visible round about the patches, owing to the scarcity of retinal pigment. A similar coloboma to these is to be seen immediately at the foot of the large coloboma. A retinal artery crosses its outer end.

Case 12 (Fig. 14, Plate VIII.).—Same patient.

Left eye.—Disc round, rosy-pink, seen best with + 0.5 D S. Vessels normal. In the triangle formed by the two temporal arteries, as they diverge from the centre to the apparent outer margin of the disc, the disc seems to be wanting, and its place is occupied by a chalk-white background covered with several choroidal vessels, and over all three curved, rod-like patches of retinal pigment. The appearance of this part of the disc closely resembles the corresponding portion of the disc in the opposite eye. It consists of choroid and retina, and differs entirely from the rest of the disc.

Half a P D to the outer side of the disc is the commencement of a large, irregular-shaped coloboma, 4 P D's × $2\frac{1}{4}$ P D's, which is exceedingly well defined and sharply bordered. Its long axis (4 P D's) is directed obliquely upwards and outwards. Half a P D external to this patch, and in a line with its long axis, is a second patch, 3 P D's square. It gives one the idea as being part of the former patch, a bridge of normal retino-choroidal tissue separating it off. The disc is seen best with + 1.5 D, the white floor of the first-mentioned white patch with — 1 D (direct), the base of the outermost (square) patch with — 2 D.

Inner coloboma.—On the inner curved border of the coloboma two clusters of pigment may be observed passing for a short distance across half of the coloboma. These are evidently the remains of trabeculæ, which stretched across the ectasia to the pigment network covering the nævus. The layer of pigment on the inner side of the gap is separated from the tissue outside by a whitish border of thinned choroid. A few choroidal veins may be seen crossing the patch to enter the nævus ; a branch from each outer retinal artery passes across both patches.

The outer half of the coloboma is chiefly occupied by a large nævus, which is made up of four thick processes of nævoid tissue, which spring from the opposite sides of the coloboma, and unite in the centre to form a dense red mass of choroidal vessels. The limbs with the outer border enclose three egg-shaped gaps, similar to the big inner patch above described, only more covered with pigment. A dense network of retinal pigment spreads itself over the whole of this half of the patch, forming the characteristic trabeculæ which pass in festoons towards the centre network.

The outer coloboma closely resembles the inner half of the coloboma described above. It is surrounded by a well-defined, free edge of retina and choroid, but the pigment is scanty. There is an attempt at the formation of a choroidal nævus at the outer half of the patch, but it is not covered by any pigment. A few choroidal vessels appear in the upper half, while five or six branches of the retinal arteries pass onwards across the gap. *It is curious that, although the arteries pass freely across, the veins do not do so, nor have I noticed any case as yet in which a retinal vein has crossed an extra-papillary coloboma.* It is true the veins do so in Fig. 12, but that includes the disc.

The sclerotic floor of this patch is seen with — 2 D, and the edge with + 1.5 D, making $3\frac{1}{2}$ D between the two levels.

V. F. : Contracted for white and colors to outer side. A large marginal gap contracts the field in the upper nasal quarter, corresponding very roughly only with the coloboma. There is no scotoma. See Chart No. 8.

I cannot conclude this article without acknowledging my indebtedness to Mr. Lawson and Mr. Tweedy for their great kindness throughout, and for the liberal way in which they have permitted me to make use of their cases.

Fig 1

W Heard Del

Fig 2

Imp 6 Dyer Bridge London

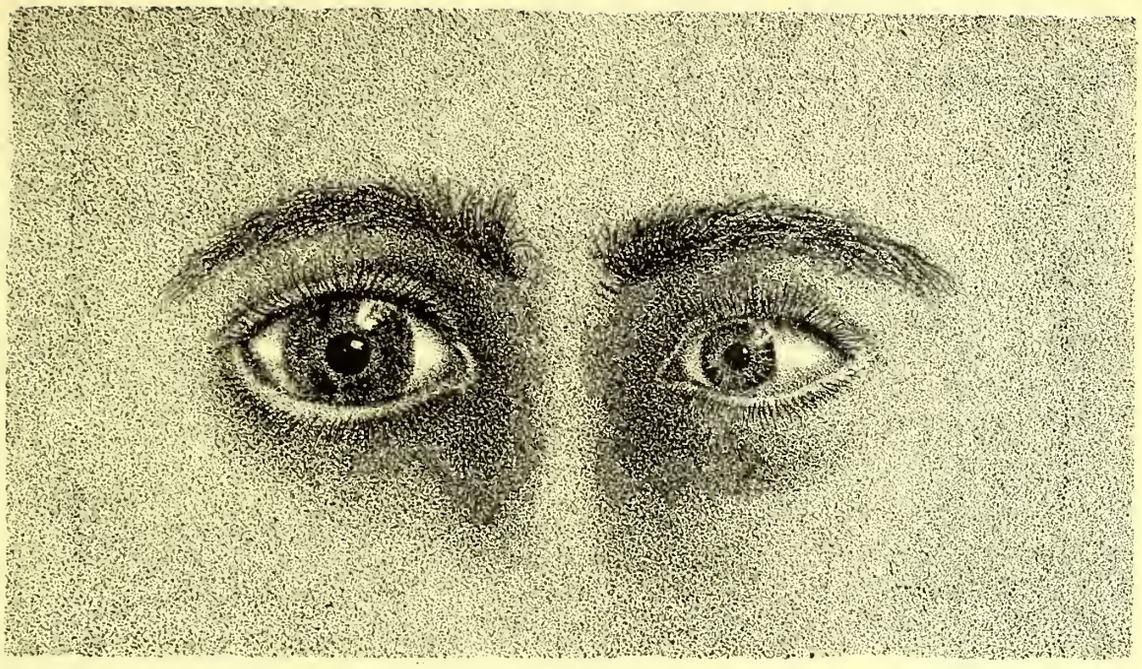

Fig 3

Fig 4

Fig 5.

A W Head del. Fig 6. Imp 5 Dyson Bldgs London

Fig 7

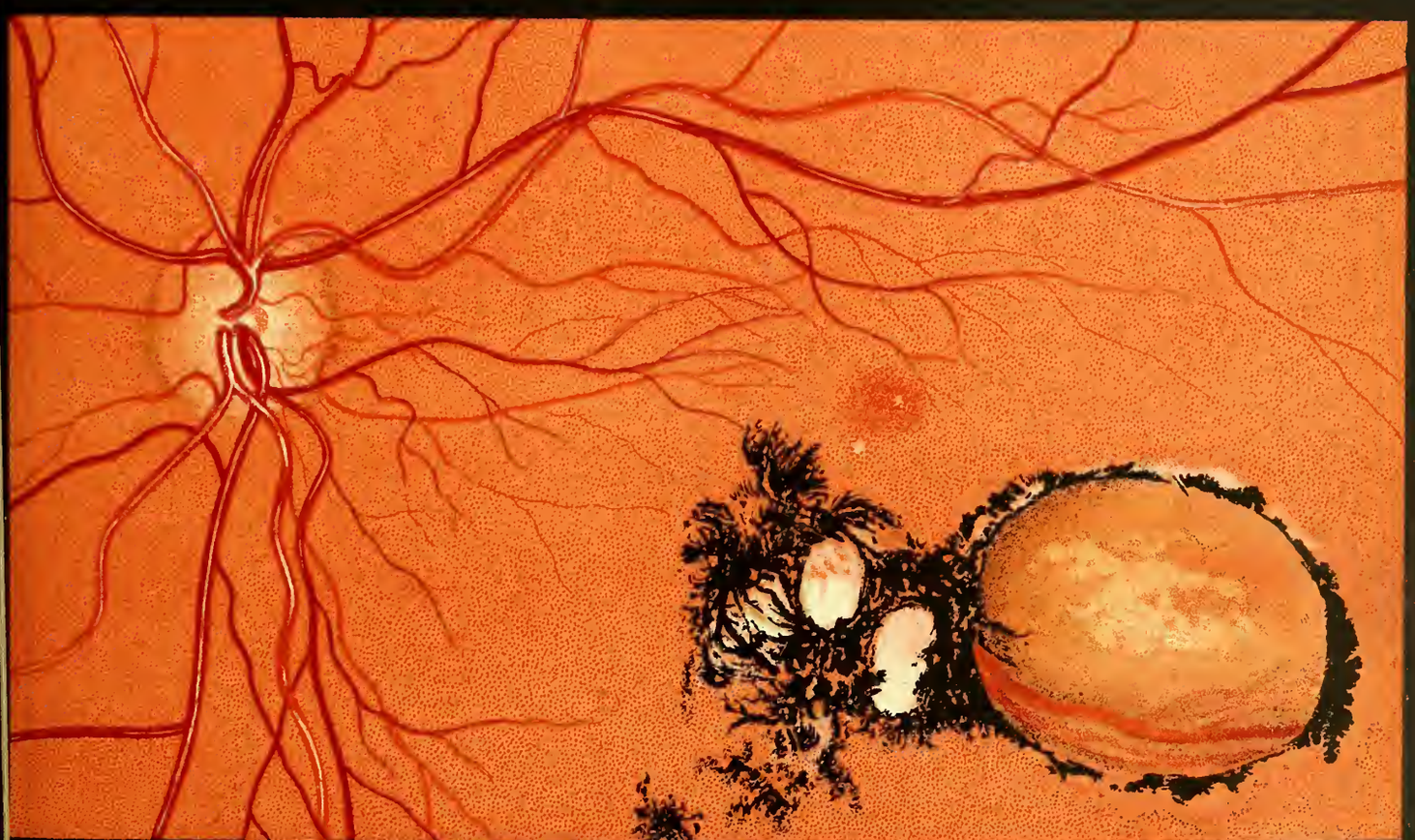

Fig 8

Fig 9

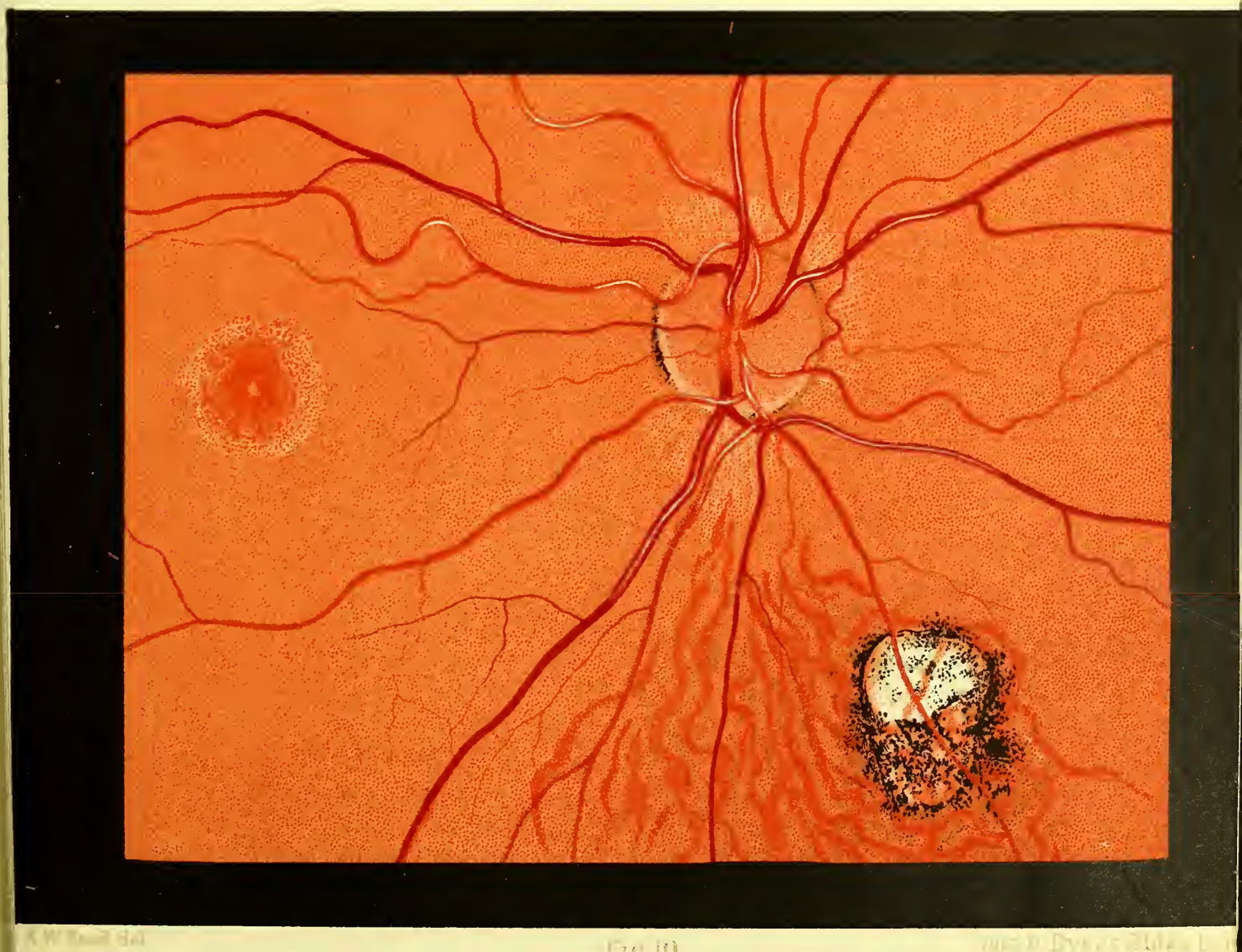

Fig 10

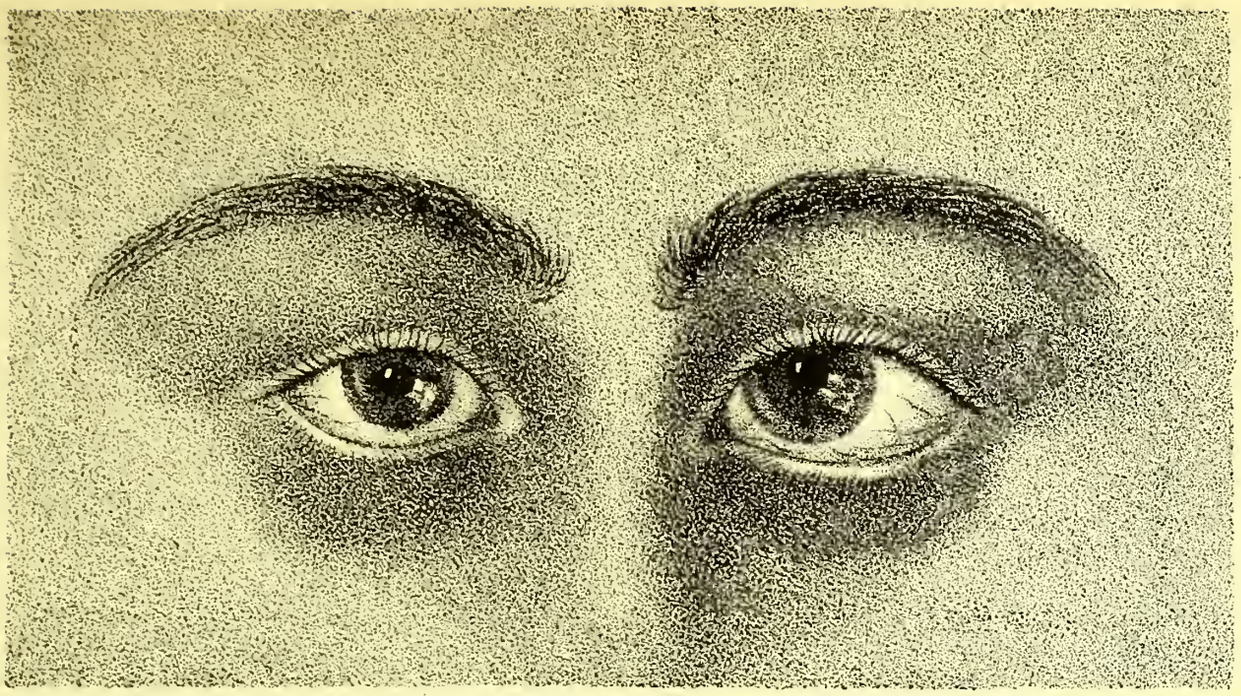

Fig 11

Fig 12

Fig 13

A.W.Head del